Using human induced pluripotent stem cells to improve our understanding of Idiopathic Pulmonary Fibrosis

Grant Award Details

Using human induced pluripotent stem cells to improve our understanding of Idiopathic Pulmonary Fibrosis

Grant Type: Tissue Collection for Disease Modeling

Grant Number: IT1-06570

Project Objective: The project objective is to collect blood samples and relevant clinical data from patients with idiopathic pulmonary fibrosis for use in generating iPSC lines as part of CIRM's iPSC Initiative.

Investigator:

Name:	Brigitte Gomperts
Institution:	University of California, Los Angeles
Type:	PI

Disease Focus: Respiratory Disorders

Award Value: $811,231

Status: Closed

Progress Reports

Reporting Period: Year 1

View Report

Reporting Period: Year 2

View Report

Reporting Period: NCE Year 3

View Report

Grant Application Details

Application Title:	Using human induced pluripotent stem cells to improve our understanding of Idiopathic Pulmonary Fibrosis
Public Abstract:	Idiopathic Pulmonary Fibrosis (IPF) is a progressive and generally fatal disease that causes scarring of the lungs and therefore an inability to breathe. Its true prevalence is unknown, as it may go unrecognized for many years, but it is generally thought to affect more than 200,000 people in the USA and is five times more common than cystic fibrosis or amyotrophic lateral sclerosis. The mortality from the disease is very high with about two-thirds of patients dying within five years of diagnosis. The cause and reasons for progression of disease are unknown, but are likely complex and multifactorial, involving genetic predisposition and environmental exposures. A small percentage of cases run in families. Our lack of therapies and understanding of IPF may in large part be due to the fact that there are no good models of the disease. Therefore, the potential development of a model of IPF from iPSC derived from IPF patients would be a major advance for the field and has the potential for drug and pathway discovery that would make a huge impact on patients' lives. [REDACTED] has one of the largest IPF clinics on the west coast and has a Clinical Trials Group and a registry for IPF. Here we propose to recruit IPF patients from the IPF clinic to donate a blood sample for iPSC derivation. We will also track their demographic and medical data. These iPSCs will then be used for disease modeling of IPF followed by high throughput drug discovery testing to identify therapies for IPF.
Statement of Benefit to California:	Idiopathic pulmonary fibrosis (IPF) is a devastating fatal lung disease that usually occurs in the 7th decade of life. The disease causes scarring of the lungs that make it impossible to breath. The prevalence of IPF is on the rise and expected to double in the next 20 years as the U.S. population continues to age. The prevalence worldwide in the population that is greater than 65 years of age is predicted to be around 125 IPF cases per 100,000 people. California, the most populous state, is also the state with the largest number of people 65 years of age and over (3.6 million people in the year 2000) and therefore the prevalence of IPF in the USA is highest in California with over 5,000 cases. By virtue of the severity of this disease, patients will almost always progress to end stage lung failure and the only therapy available is a lung transplant. In addition to the suffering and disability that IPF causes, the costs associated with caring for patients with IPF, including lung transplant costs, are extremely large. For example a single lung transplant costs roughly $400,000 and this doesn't include all the follow up care needed post-transplant or any complications. Improving our understanding of and identifying therapies for IPF will therefore have a major impact on patients in California with IPF. Firstly, it will reduce much pain and suffering and secondly it will reduce costs and free up donor lungs, that are in short supply, for other end stage lung disease patients.